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HERPETIC INFLAMMATIONS OF THE GENICULATE GANGLION. A NEW SYNDROME AND ITS AURAL COMPLICATIONS.[1]

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(*With two illustrations on Text-plate II.*)

INTRODUCTION.

THE subject of this communication was presented at a meeting of the American Neurological Association held in Boston, June, 1906. A further presentation was made at the conjoint meeting of the New York and Philadelphia Neurological Societies held in New York, March, 1907. These dealt in detail with the neurological aspects of the subject. In the present communication the otological features will be emphasized more particularly, especially the otalgia, the herpes zoster of the concha and external auditory canal, and the symptoms of Ménière's disease which represent important aural complications of the affection.

Briefly, the syndrome is dependent upon a specific herpetic inflammation of the geniculate ganglion, situated on the facial nerve in the depths of the internal auditory canal in the entrance to the aqueduct of Fallopius. The simplest expression of this inflammation is to be found in the herpes zoster of the tympanum, auditory canal, and the concha (*this representing the zoster zone for the geniculate ganglion*). Because of the proximity of the facial nerve and the terminations of the auditory nerve, neural complications are not infrequent: peripheral facial palsy, tinnitus aurium, deafness, and symptoms of Ménière's disease. The pathology of the affection does not differ from that of true herpes zoster, or zona. This, as has been shown by the elaborate pathological studies of Head and Campbell, depends upon a specific inflammation of the posterior spinal ganglia. While one ganglion is usually the seat of the chief manifestation of the disease, the neighboring and adjacent ganglia may also show slight inflammatory changes. Evidence of this multiplicity of lesions is shown clinically by the occurrence of double bilateral and, rarely, triple forms of zona, which are especially frequent in the region of the face, head, and neck.

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[1] Read before the Section on Otology of the New York Academy of Medicine, April, 1907.

The facial nerve is a mixed nerve. Its sensory ganglion is the geniculate; its afferent root is the nerve of Wrisberg; its efferent, the facial nerve proper. The peripheral divisions of the geniculate ganglion are the great and small superficial petrosal nerves and the external petrosal nerve, the latter passing to the carotid plexus of the sympathetic; the former, to Meckel's and the otic ganglia respectively. Furthermore, both the greater and lesser superficial petrosal nerves participate in the formation of the tympanic plexus through the great and small deep petrosal nerves. (See Fig. 1.) As the geniculate ganglion contains cells of the spinal ganglion type, it, like the Gasserian ganglion, may properly be brought within the realm of the specific inflammation of herpes zoster.

The ganglionic representations of the cephalic extremity of the body roughly outlined are as follows: The Gasserian ganglion for the face, the geniculate ganglion for the concha and auditory canal, the second, third, and fourth cervical ganglia for the occiput and neck,—any zoster inflammation of these giving rise respectively to herpes facialis, herpes auricularis, and herpes occipito-collaris. As the tendency of the specific inflammation of zona is to produce inflammatory changes in a series of ganglia, although its chief manifestation is in a single ganglion, it would follow that when the chief focus is in the Gasserian ganglion, an inflammatory reaction might also occur in other ganglia of this series (the geniculate, second and third cervical ganglia).

In this manner I would explain that group of cases with facial palsy and acoustic symptoms in which the eruption is distributed over the face or neck, the zone for the geniculate ganglion itself remaining free. I have collected sixty-one cases of true herpes zoster including five personal observations presenting a peripheral facial palsy, in which the eruption was distributed over the face, neck, or ear. Of this number, twenty showed also an involvement of the auditory nerve. The distribution of the eruption in these cases was as follows:

Herpes occipito-collaris	32	cases.
Herpes auricularis	13	"
Herpes facialis	12	"
Herpes occipito-collaris and auricularis	3	"
Herpes facialis and occipito-collaris	1	case.

The herpetic inflammation of the geniculate presents the following clinical types:

1. Herpes auricularis.
2. Herpes auricularis, facialis, or occipito-collaris, with facial palsy.
3. Herpes auricularis, facialis, or occipito-collaris, with facial palsy and hypoacousis.
4. Herpes auricularis, facialis, or occipito-collaris, with facial palsy, deafness, and symptoms of Ménière's disease.

HERPES AURICULARIS, OR HERPES OTICUS.—This manifestation of herpes zoster has been recognized by all systematic writers. It has, however, always been referred to the trigeminus nerve or ganglion, or to the cervical ganglia or nerves. Up to the time of the publication of my paper, the only recognized localization of an herpetic inflammation on a cranial nerve ganglion was the Gasserian. While there is no question that the eruption may involve the anterior and posterior portions of the auricle in cervical and facial herpes, there is a well-defined clinical type dependent upon a geniculate inflammation in which the eruption is distributed over the concha, external auditory canal, and the tympanic membrane. I would therefore restrict the use of the terms "herpes oticus" and "herpes auris" to this distribution.

As with herpes zoster in general, mild prodromal symptoms not infrequently precede the onset of the pain. In herpes auris, the preherpetic pains are referred to the depths of the ear, external auditory canal, and the auricle. These may reach an extreme degree of severity. In a few cases the tympanic membrane itself has been incised to relieve a supposed middle-ear inflammation. In severe cases not only are pains felt in the interior of the ear but they radiate also in the three branches of the fifth nerve, and to the mastoid, occipital, and cervical regions. In many cases the auricle becomes red and swollen, and quite tender, assuming an erysipelatous or frost-bitten appearance; the external auditory canal may be practically occluded by the swollen tissues, thereby inducing a mechanical interference with hearing which must not be confused with the deafness resulting from true lesions of the auditory nerve, which will be described later.

Usually on the third or fourth day the characteristic eruption makes its appearance, the abundance of the vesicles varying greatly in different cases. In rare instances the vesicles have been confined to the tympanic membrane, with all the other manifestations of true herpes zoster.[1] In one of my own cases with facial palsy and hypoacousis, only two very small patches were present, one at the inferior border of the external meatus, the other beneath the anthelix.

From an analysis of thirteen cases of herpes oticus, I would confine the zoster zone to the tympanum, external auditory canal and meatus, the concha, tragus, and antitragus; also an adjacent marginal area which coincides with the tendency of these zones to overlap.

As bilateral herpes zoster of the Gasserian ganglion is occasionally observed, it would seem probable that the geniculate ganglia might also be the seat of a bilateral involvement. Such a case is recorded by Chavanne.[2]

The eruption follows the usual course of zoster, all trace, save a few pigmented scars, disappearing in the course of a fortnight. Post-herpetic otalgias in this group of cases are often very severe and persistent, especially in the old.

HERPES AURICULARIS, FACIALIS, OR OCCIPITO-COLLARIS WITH FACIAL PALSY.—In this group the inflammation has extended to the trunk of the facial nerve. Its confinement within a narrow bony canal may also by pressure play a rôle in the production of this symptom. The palsy may appear with the eruption, or it may be delayed a week or even longer. As its onset is often insidious, it is frequently overlooked, so that too much stress should not be laid upon the patient's statements as to the time of its appearance. As the occurrence of repeated crops of vesicles are not infrequent in zoster, it is possible that a succession of inflammatory changes may be taking place within the affected ganglion. This might in some cases explain the length of time intervening between the appearance of the eruption and the palsy. The palsy in all of the sixty-one cases was complete, involving all branches of the nerve.

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[1] Szenes, "Herpes Zoster des Trommelfelles," *Internat. Centralbl. für Ohrenheilk.*, i., 1902.

[2] "Zona bilaterale, isole de l'oreille," *Annal. des mal. de l'oreille*, 1906, p. 639.

A rather striking peculiarity is the not infrequent evanescence of this symptom, in some cases lasting only a few days, and many of them clearing up within a month. More severe types of the paralyses, however, occur with reactions of degeneration, and leaving permanent weakness and contractures. In a very large proportion of cases the sense of taste was affected on the anterior two-thirds of the tongue, which is readily explained by the involvement of the chorda-tympani nerve at the geniculate level.

As has already been remarked, facial palsies may occur with herpes occipito-collaris, and herpes facialis, as well as with herpes auricularis. This I would explain by the simultaneous occurrence of inflammatory changes in a series of ganglia.

HERPES AURICULARIS, FACIALIS, OR OCCIPITO–COLLARIS, WITH FACIAL PALSY AND HYPOACOUSIS.—In this group, in addition to the herpetic manifestations and the facial palsy, there is hypoacousis. The presence of auditory symptoms is to be explained by the extension of the products of inflammation to the acoustic nerve. If this is transitory and slight, it is the result of inflammatory œdema only; if more severe, the nerve trunk is probably directly invaded by the inflammatory process.

HERPES AURICULARIS, FACIALIS, OR OCCIPITO-COLLARIS, WITH FACIAL PALSY, DEAFNESS, AND SYMPTOMS OF MÉNIÈRE'S DISEASE.—This is the severest and most interesting manifestation of the disease. Here, in addition to the facial palsy and deafness, there are present the classical symptoms of Ménière's syndrome,—tinnitus aurium, disturbances of the equilibrium, nystagmus, vomiting, and labyrinthine vertigo. Of the twenty cases which presented symptoms of involvement of the auditory nerve, nine showed hypoacousis only. In the other eleven the symptoms of Ménière's disease were present.

The distribution of the eruption in these cases was as follows:

Herpes occipito-collaris	9	cases
Herpes auricularis	7	"
Herpes facialis	4	"

Before leaving this subject I would call attention to the occasional presence of auditory symptoms in cases of herpes about the face and neck in which facial palsy was absent. Gruber[1] noted in his cases of herpes auris that there were tinnitus and diminished hearing, and referred these to a probable reflex involvement of the auditory nerve. The same peculiarity was observed by Vail[2] in his case of herpes auris. As facial palsy in these cases is often very transient, it may possibly have been overlooked, for if, as I infer, the auditory symptoms are dependent upon an extension of the inflammation from the geniculate, one would hardly expect to find them present without facial palsy, as the inflammation must traverse the trunk of the seventh to reach that of the eighth.

I also find that in some cases of herpes facialis auditory symptoms were present without facial palsy (Cassé[3] and Burin-Desrozier[4]), so the thought naturally suggests itself that the ganglia of the acoustic nerve may also be brought within the realm of the specific infection of herpes. (See Fig. 2.)

It will be recalled in this relation that the ganglion acousticum is an outgrowth of the so-called neural ridge, from which the Gasserian, geniculate, and posterior spinal ganglia take their origin. In the course of development the cells of the geniculate ganglion assume the spinal unipolar type, whereas those of the acoustic ganglia (ganglion of Corti, and ganglion of Scarpa) retain their primitive bipolar character. But the extreme severity and persistence of the acoustic symptoms in some of these cases, and the occurrence of acoustic symptoms without facial palsy, and the origin of these cells in the neural ridge, seem to me strong evidence that herpetic inflammation of the acoustic ganglia may occur in the cephalic forms of zona.

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[1] "Herpes auricularis," *Monats. f. Ohrenheilk.*, 1875, No. 5.

[2] "Herpes zoster auris," *Jour. Laryngol. and Rhinol.*, 1907.

[3] "Zona de la troisième branche du trijumeau," *Thèse de Paris* 1901.

[4] "Zona de la deuxième branche du trijumeau," *Thèse de Paris*, 1904.

DIAGNOSIS.—The diagnosis of these cases is not difficult. The prodromal symptoms, the intense neuralgic character of the pain (pre- and post-herpetic neuralgia and otalgia), with the appearance of the characteristic eruption, should separate this from all other affections of the ear; and yet I believe that many of the cases are not recognized and properly classified. The area of the eruption is often so small, and the redness, swelling, and tenderness of the auricle so great, that many cases, I have no doubt, are regarded as diffuse inflammations of the auricle, perichondritis, frost-bite, and the like. In the later stages, with itching and scaling, the affection might be confused with eczema of the auricle.

That the condition is frequently overlooked would seem to be confirmed by hospital statistics. That the affection is not a common one is shown by Gruber's series, who found five cases in 20,000 cases of ear disease. To ascertain, if possible, some knowledge of the frequency of the affection in this country, I examined the annual reports of several of our large eye and ear hospitals, with the following result:

In the Manhattan Eye and Ear Hospital, during a period of ten years, with a total of 47,600 cases, the diagnosis of herpes auris was made in two instances.

In the Brooklyn Eye and Ear Hospital, during a period of five years, with a total of 15,000 cases, the diagnosis was made but once.

In the New York Eye and Ear Infirmary, one of the largest institutions of its kind, during the past twenty years, this diagnosis was recorded only six times.

In the Massachusetts Eye and Ear Infirmary, however, during a period of ten years, with a total of 65,000 cases, the diagnosis was recorded thirty-three times.

In these reports it is not stated whether the zoster was primary or secondary, but the infrequency with which the diagnosis was made is worthy of note.

The recognition of those types of cases in which the facial nerve and acoustic complications are present should not give rise to difficulty, unless seen after the disappearance of the eruption; even then a retrospective diagnosis might be possible from the character and severity of the initial herpetic pains, and the presence of pigmented scars. I would particularly emphasize the importance of a careful examination of the tympanum, auditory canal, and the recesses of the auricle, as the small patches of vesicles may be concealed and overlooked. The facial palsy occurs with, or follows soon after, the eruption,—auditory symptoms, if present, manifesting themselves about the same time. It should also be borne in mind that symptomatic herpes zoster of the auricle may complicate otitis media and tumors in this region.

Prognosis.—The prognosis in these cases is generally very favorable. The exceptions are the occasional persistence of tinnitus and deafness in the Ménière's type. Residual symptoms of facial palsy may also persist. Very rarely in old people, prolonged and intractable otalgias may continue for a considerable time.

It would seem advisable, as the symptoms are dependent upon hemorrhagic inflammations of the neural structures in the depths of the auditory canal, to leech the mastoid region in all cases as soon as the diagnosis is made, in the hope that this may prevent serious inflammatory invasion of the nerve trunks.

Concluding remarks.—The syndrome which I have outlined, depends for its characteristic features upon the relation of the geniculate ganglion to the seventh and eighth nerves, which are enclosed in a common sheath and course within a narrow bony canal. The syndrome is composed of three distinct groups of cases long recognized and the subject of much discussion. The clinical relation of these groups to one another and their common pathology has not been recognized. In the group with facial palsy, the paralysis was regarded as a rheumatic or toxic neuritis. The same interpretation was given to the group with auditory symptoms. Some idea of the confusion attending the classification of this latter group may be obtained from a perusal of the following list of titles: Polyneuritis cerebralis Ménièreformis (Frankl-Hochwart, 1899, and Berger, 1905); Ueber ein Fall von gleichseitigen, akut aufgetretene Erkrankung des Acusticus, Facialis und Trigeminus (Kaufmann, 1896); Zur Lehre von der peripherischen Facialis Lähmung (Hoffmann, 1899); Beitrag zur Casuistik der multiplen Hirnnerven Erkrankung (Hammerschlag, 1898); Herpes Zoster Oticus (Körner, 1904); Trouble auditive dans le zona (Lannois, 1904).

NOTE—For a full bibliography and discussion, see author's article "On Herpetic Inflammations of the Geniculate Ganglion," *Journal of Nervous and Mental Diseases*, February, 1907; also The *Transactions* of the American Neurological Association, 1906, page 184; and the "Transactions of the Meeting of the New York Neurological Society" held March, 1907, *Journal of Nervous and Mental Diseases.*

EXPLANATION OF TEXT-PLATE.

FIGURE 1. The geniculate ganglion (intumescentia ganglioformis); its relation to the facial and auditory nerves and its communications (Cunningham's Anatomy).

VII., Facial nerve.
P. I., Pars intermedia.
VIII., Auditory nerve.
Aq. Fal., Aqueduct of Fallopius.
G, G, Geniculate ganglion.

ILLUSTRATING DR. HUNT'S ARTICLE ON HERPETIC INFLAMMATIONS OF THE GENICULATE GANGLION.

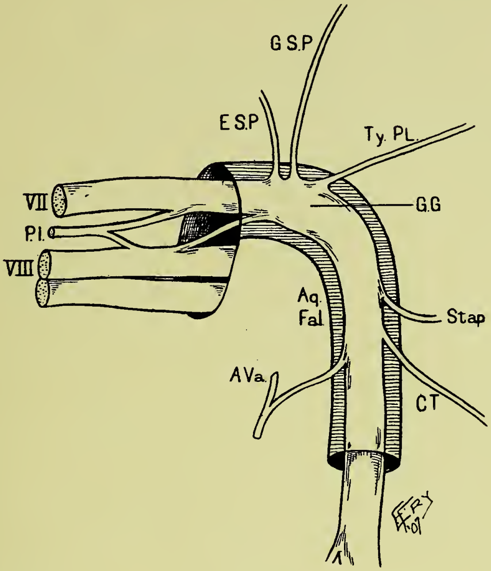

FIG. 1.—The relations of the geniculate ganglion.

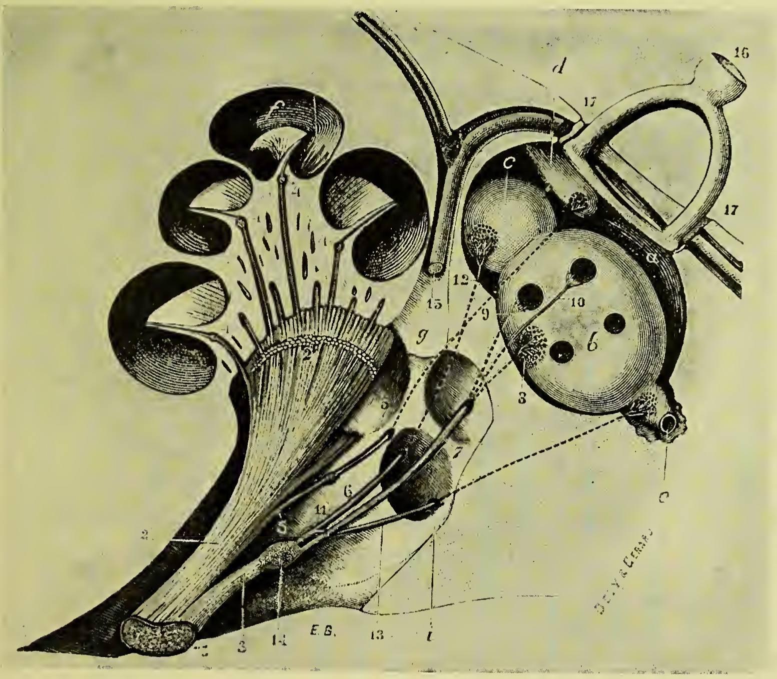

FIG. 2.—The terminations of the auditory nerve.

E. S. P., External superficial petrosal nerve.
G. S. P., Great superficial petrosal nerve.
Ty. Pl., Branch to the tympanic plexus.

FIGURE 2. The ganglia and terminations of the auditory nerve (Testut's Anatomy).

1. Auditory nerve.
2. Cochlear branch.
3. Vestibular branch.
4. Ganglion of Corti.
6. Ganglion of Boettcher.
14. Ganglion of Scarpa.
15. Facial nerve.